

Primary ciliary dyskinesia

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- Older name: immotile cilia syndrome; OMIM: 244400 (<http://omim.org/entry/244400>)
- It is a generalized disorder of motility of cilia and flagella of all cells, inherited in an autosomal recessive manner.
 - The movement is either completely absent or is uncoordinated.
 - It was originally described as part of Kartagener's syndrome (bronchiectasis, sinusitis, situs viscerum inversus).

Clinical picture

- Impaired self-cleaning ability of the epithelium, productive expectoration and bronchiectasis occur.
- Other symptoms: sinusitis, chronic rhinitis, rhinorrhea, nasal polyps, recurrent otitis, conduction disorder hearing loss, tympanic perforation, DC obstruction, recurrent pneumonia, infertility.
- Situs viscerum inversus only occurs in about 50% of cases.
- Diagnosis: complicated confirmation and it is necessary to know the result of electron microscopy and high-speed video microscopy (HSVM).

Therapy

- Symptomatic, similar to other bronchiectasis.

Links

Related articles

- Chronic lung diseases

References

- BENEŠ, Jiří. *Studijní materiály* [online]. [cit. 2010]. <<http://jirben.wz.cz>>.

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